Medline

Creutzfeldt-Jakob disease and cerebral amyloid angiopathy.

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Abstract

An 83-year-old female with no personal or familial neurological history developed progressive gait and speech disturbance and left motor deficit. She suffered intractable seizures and died 3 months after the onset of neurological signs. Neuropathology showed severe spongiosis and gliosis in the cortex and basal ganglia, and diffuse cerebral amyloid angiopathy. Immunostaining for prion protein (PrP) showed intense PrP positivity in areas of confluent spongiosis and some granular staining in astrocytes. The cortical vessel walls stained positively for beta/A4 amyloid but not for PrP amyloid. Both types of amyloid were only observed in pericapillary parenchyma, in areas with severe spongiosis. There were only a few tangles and neuritic plaques in the temporal cortex; amyloid plaques were not present either by silver stains or immunostains. There was neither arteriopathic leukoencephalopathy nor cerebral hemorrhage. Immunoblot analysis of brain extracts revealed an abnormal proteinase K-resistant isoform of PrP. Association of Creutzfeldt-Jakob disease and Cerebral amyloid angiopathy in the absence of Alzheimer changes in unusual. The association of PrP and beta/A4 amyloid deposits could have been fortuitous in an 83-year-old patient. An etiopathogenic relationship between beta/A4 amyloid deposition and PrP accumulation may also be considered.

MeSH

Aged; Aged, 80 and over; Amyloid beta-Protein; Brain; Case Report; Cerebral Amyloid Angiopathy; Creutzfeldt-Jakob Syndrome; Female; Human; Immunoenzyme Techniques; Prions; Support, Non-U.S. Gov't

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711+1, 1717-1 and 3659 as disclosed herein, carrier screening and prenatal diagnosis can be carried out as follows.

The high risk population for cystic fibrosis is Caucasians. For example, each Caucasian woman and/or man of child-bearing age would be screened to determine if she or he was a carrier (approximately a 5% probability for each individual). If both are carriers, they are a couple at risk for a cystic fibrosis child. Each child of the at risk couple has a 25% chance of being affected with cystic fibrosis. The procedure for determining carrier status using the probes disclosed herein is as follows.

For purposes of brevity, the discussion on screening by use of one of the selected mutations is directed to the I507 mutation. It is understood that screening can also be accomplished using one of the other mutations or using several of the mutations in a screening process or mutation detection process of this section on CF screening involving DNA diagnosis and mutation detection.

One major application of the DNA sequence information of the normal and 507 mutant CF gene is in the area of genetic testing, carrier detection and prenatal diagnosis. Individuals carrying mutations in the CF gene (disease carrier or patients) may be detected at the DNA level with the use of a variety of techniques. The genomic DNA used for the diagnosis may be obtained from body cells, such as those present in peripheral blood, urine, saliva, tissue biopsy, surgical specimen and autopsy material. The DNA may be used directly for detection of specific sequence or may be amplified enzymatically in vitro by using PCR [Saiki et al. Science 230: 1350-1353, (1985), Saiki et al. Nature 324: 163-166 (1986)] prior to analysis. RNA or its cDNA form may also be used for the same purpose. Recent reviews of this subject have been presented by Caskey, [Science 236: